CLAIMS

1. Transgenic non-human animal, preferably a knock-in mouse, having a missense mutation in the α4- or β2-subunit of the neuronal nicotinic acetylcholine receptor (nAChr).

2. Animal of claim 1, having the missense mutation V287L or V287M in the gene for the β2-subunit.

3. Animal of claim 1, having the missense mutation 259-260ins, S252L, 766ins3 or T265I in α4-subunit of the nAChr receptor.

4. Animal of any one of claims 1 to 3 containing the missense mutation homozygously.

5. Animal of any one of claims 1 to 3 containing the missense mutation heterozygously.

6. Targeting vector containing the following components operatively linked:

- the genomic and/or cDNA sequence for a subunit of the, preferably human or murine, nicotinic acetylcholine receptor (nAChr) having a missense mutation in the α4- or β2-subunit, or a part of said subunit, wherein said part has at least the missense mutation in the α4- or β2-subunit.
- a selectable marker gene, and
- optionally 2 recognition sequences for a recombinase, which flank the marker gene.

7. Targeting vector of claim 6, wherein the selectable marker is an antibiotic resistance gene.

8. Targeting vector of claim 6 or 7, wherein the recognition sequences are each loxP.

9. Targeting vector of one or more of claims 6 to 8, wherein the β2-subunit has the missense mutation V287L or V287M.

10. Targeting vector of one or more of claims 6 to 8, wherein the α4-subunit has the missense mutation 259-260ins, S252L, 766ins3 or T265I.

11. Stem cell, preferably murine embryonic stem cell, containing a vector of one or more of claims 6 to 10.

12. Screening method for the identification of compounds for the treatment of the human epilepsy syndrome, particularly familiar nocturnal frontal lobe epilepsy (ADNFLE), comprising the following steps:

a) providing an animal of any one of claims 1 to 5 and providing test compounds,

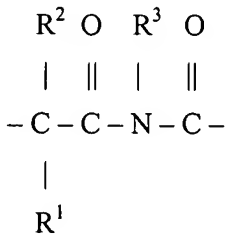
b) administration of the test compounds to the animal,

c) selection of a test compound resulting in alleviation or elimination of the symptoms of the epilepsy syndrome in the animal, and

d) optionally repeating the steps a) to c) with a suitably modified form of the test compound chosen in c).

13. Screening method of claim 12, wherein the test compounds are selected from the following groups:

- barbiturates, oxazolidindiones and succinimides and further groups having the following grouping as a common structural element:



wherein R^1 and R^2 are alkyl or aryl residues and R^3 is H or an alkyl residue,

or

- derivatives of benzodiazepines, sultiam, Carbamazepin and valproic acid.

14. Compound for the treatment of the human epilepsy syndrome, preferably of ADNFLE, which has been identified by the method of claim 12 or 13.

15. Pharmaceutical composition having a therapeutically effective dose of one or more compounds of one or more of claims 12 to 14 and a pharmaceutically acceptable carrier.

16. Use of the composition of claim 15 for the treatment of the human epilepsy syndrome, preferably ADNFLE.